Nodular Fasciitis: A Tumor with Misnomer!

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Abstract
Nodular fasciitis is a peculiar tumor in its clinical presentation and cytological appearance, owing to its similarity with soft tissue sarcoma. It is a rare benign neoplasm most commonly occurring in the upper extremity. Meanwhile it is self-limiting reactive process mimicking malignant lesion composed of Fibroblasts and Myoblasts. Here we are reporting two cases in the hand, with varied clinical presentation, were treated with complete surgical excision and pathologically confirmed to be Nodular Fasciitis. The term Nodular Fasciitis although pathologically is indicative of an inflammatory process, clinically it seems to be misleading since “Fasciitis” usually denotes an acute serious soft tissue infection. As per literature, this benign tumor is often described as pseudo-sarcomatous, yet its title gives no hint of such a concern. After dealing with the presented cases and the literature review it is evident that, diagnosing this specific lesion on basis of clinical picture, MRI or fine needle aspiration is very unlikely. Most cases will end up having surgical excision due to its frequently alarming presentation.

Introduction
Nodular fasciitis (Nod. Fasciitis) was originally described by Konwaler and Weiss in 1955 as pseudo-sarcomatous fasciitis, pseudo-sarcomatous fibromatosis or Infiltrative fasciitis. Clinically it would present with relatively short history, rapid growth, palpable soft tissue tumor with occasional pain [1, 2]. Pathologically Nod. Fasciitis is an encapsulated lesion that is typically well demarcated from the surrounding uninvolved tissue but may be focally infiltrative [3]. This tumor sometimes gives mimicking histopathological picture of spindle cell sarcoma, fibrous histiocytoma, pleomorphic adenoma, proliferative fasciitis and benign nerve sheath tumor.

Nod. Fasciitis is a rare benign neoplasm most commonly effecting the soft tissue of Upper extremity followed by Trunk, Head & Neck. It is commonly seen in the 4th(fourth) decade with no gender predilection [4, 5]. Its self-limiting reactive pseudosarcomatous process composed of fibroblast and myofibroblast. The pathogenesis of Nod. fasciitis is unknown. There is an association with the previous trauma in 5-10 % of the cases. That is in fact related to a hyper response mechanism triggering an increase in mitotic activity in certain pre-disposed individuals[6].

Non-surgical management with close observation alone as therapeutic modality although mentioned earlier by some authors, it is not generally recommended. This is due to risk of incorrect diagnosis and the existence of potentially a malignant lesion. If diagnosed on clinical and radiological basis, N. Fasciitis should be surgically excised to confirm diagnosis and to ensure complete cure. The chances of local recurrence are almost nil [7]. Here we are presenting the following two cases

Case 1: This was a 51 years old right-hand dominant lady sent to us with recurrent soft tissue tumor on her left-hand little finger of one-year duration, she was otherwise healthy. There was no history of injury. The patient had it initially excised at another hospital 6 months back, with early recurrence of the tumor, no pathology report was available. The patient indicated, the tumor has been growing significantly for last few weeks.

On clinical examination she had a granulomatous swelling at the terminal phalanx of the left hand 5th digit. It was around 2.5 x 2 cm in dimension bright red mass, non-mobile (Figure 1).

Case 2: This was a 42 years old lady with history of previous right breast implant removal due to infection. She presented with lump in the left wrist of 6 months duration.

Case 2 upon presentation, 5th digit volar aspect, a fingertip tumor of one-year duration, with history of recurrence post excision and accelerated growth. It is firm, immobile.
Laboratory tests for inflammatory markers and rheumatological conditions were negative. Base line X-Rays showed exostosis and bone involvement (Figure 2).

![Figure 2](image)

**Figure 2**
Plain x-ray, soft tissue shadow and involvement of terminal phalanx with exostosis.

As the lesion was evident on the surface an incisional biopsy was done and it came as Nod. Fasciitis. Interestingly the tumor grew significantly over the course of 2 weeks between her biopsy to definitive excision (Figure 3, 4). Ablation of the terminal phalanx was done with appropriate stump reconstruction. Final histopathological diagnosis was the same. Patient was followed close to 6 months with complete healing and cure.

![Figure 3](image)

**Figure 3**
Case 1 cont’d. Significant growth noted only within 2 weeks following an incisional biopsy

![Figure 4](image)

**Figure 4**
Dorsal view, tumor is extending to nail bed. Prior to definitive excision and ablation of distal phalanx.
Case 2: This was a 45 years old male right hand dominant otherwise healthy school teacher. He was also referred from another institution with a soft tissue mass at his left hand 5th digit base. Patient denied any trauma preceding the presence of the swelling. He gave history of continuous, steady and gradual growth of this soft tissue mass over few months (Figure 5). There was a soft tissue swelling at the ulnar-volar aspect of the base of the little finger, left hand. This was non-tender, firm mass with restricted movement at all directions. Peripheral neurovascular status of the digit was normal.

Figure 5

Case 2, Swelling in proximal little finger of few months’ duration, it is subcutaneous, firm and non-mobile.

Figure 6

Case 2 upon exploration, tumor has pseudocapsule and attached to flexor tendon sheath.

Figure 7

Tumor been dissected off the digital nerves and vessels but it is involving the pulley’s system of tendon sheet.

Discussion

Typically, Nod. Fasciitis begins as a small relatively superficial nodule with gradual increment in size. The majority of cases manifest as solitary, painless growing tumor up to 2-3 cm within a period of 2 to 12 weeks [8].

It is considered to occur due to unusual proliferation of myofibroblasts triggered by a trivial trauma or any inflammatory process [9]. Clinical presentation is much varied and on top pathological assessment can also mimic with spindle cell sarcoma; fibromatosis; fibrous histiocytoma; proliferative fasciitis; benign nerve sheath tumor and pleomorphic adenoma. Histopathology with Immunohistochemistry helps in formulating the diagnosis of Nod. Fasciitis. It appears to be equally distributed in both genders. In order of frequency, the most common location of this benign tumor is in the upper extremity (39-54%), trunk (15-20%), lower extremity (16-18%), head and neck (7-20%) [10]. There are three main subtypes: Subcutaneous; Intramuscular; and fascial. Other less common subtypes include dermal; intravascular; cranial; and ossifying. Most lesions are subcutaneous, well circumscribed and non-encapsulated [4]. The intradermal variant can present with additional features of superficial ulceration with bleeding and they

Radiological study including MRI concluded a soft tissue tumor of about 2.5 X 2.5 cm. The tumor was seemingly encircling the flexor tendons with erosion of the cortex of the proximal phalanx of the 5th digit. Based on the clinical assessment and radiologist input, the working diagnoses included tendon sheath Giant cell tumor or Schwannoma or Fibroma. Per-operatively the tumor was found to be adherent to A1 and A2 pulleys of the flexor sheath. The entire tumor had to be taken along with the pulleys, the A2 was reconstructed (Figure 6-8). Pathology report confirmed the diagnosis of Nod. Fasciitis. Patient did well with up to 6 months follow up.

Figure 8

Following complete excision of tumor and reconstruction of A2 pulley.
are associated with pain as well [11].

Histologically, these lesions are composed predominantly of fibroblast, either plump or spindle shaped and resemble granulation tissue (as in our first case). Early in the course of the lesion myxoid histology is especially prominent. It becomes more cellular with time and older lesions tend to have a fibrous histology that may be characterized by the hyaline fibrosis. Micro cysts may also form in lesions of longer duration [12]. This histologic diversity likely accounts for the variable MRI imaging appearance of the lesions. The risk of incorrect diagnosis and the existence of potentially more life-threatening lesion may encourage excisional biopsy over observation alone as the diagnostic and therapeutic modality. The option of observation after FNA based pathology of this tumor is also recommended in literature [13]. Spontaneous resolution of nodular fasciitis within a median period of 2 weeks to 16 weeks could be awaited. However, with unusual course, longer duration of symptoms, lack of resolution within the given time frame, or atypical cytological results with FNAC, are clear indication for excisional biopsy [14].

Both of our presented cases, intraoperative findings and pathological description adapt well with the reviewed literature. Nod. Fasciitis is often misdiagnosed as a soft tissue sarcoma, which is the most important pathological differential diagnosis of this condition. Some scientist claimed 2/3 of their cases were misdiagnosed as sarcoma [15]. Making a correct diagnosis of this entity must base on the clinical features, Magnetic resonance imaging and Histopathological examination with immunohistochemistry. Although rare, we find Nod. Fasciitis to be a very interesting lesion due to its clinical similarity of other benign soft tissue tumors in some instances, but in others resemblance to more serious lesions, and as well due to its confusing name.

Hand and soft tissue surgeons often deal with a similar but much more alarming lesion, “Dermatofibrosarcoma protuberans” (DFSP) which is a low grade locally aggressive sarcoma, supposedly it is curable with adequate wide local excision unless it is neglected or mismanaged [16]. Another interesting tumor which has also been managed by closed observation historically is “Keratoacanthoma”, it may regress spontaneously over a course of 3 to 6 months, but it seems majority of surgeons nowadays would not take a chance to preclude mistaken diagnosis and missing a Squamous cell carcinoma. Unfortunately, the pathological differentiation between the two entities can be of a major challenge as well [17].

The term Nodular Fasciitis can be of source of confusion for the surgeons when read on a pathology report, since “Fasciitis “usually indicates an acute life-threatening soft tissue infection (Necrotizing Fasciitis), requiring immediate surgical treatment and supportive measures. Examples of other common misnamed lesion include “Pyogenic granuloma” presenting as recurrently bleeding granular skin lesion or tumor, however pathologically it is neither purulent nor a granulation tissue. Another one is the “Benign juvenile Melanoma”!

References